CLINICAL PROCEEDINGS

of the

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

July 1952

VOLUME VIII NUMBER 7

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CLINICAL PROCEEDINGS

OF THE CHILDRENS HOSPITAL

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A SUGGESTED MANAGEMENT OF ANTERIOR POLIOMYELITIS IN THE ACUTE STAGE

EDITORIAL

Poliomyelitis is, perhaps, the most misunderstood and mismanaged disease in childhood. Undue publicity and concern are the factors most responsible for this situation. The etiologic agents responsible for acute anterior poliomyelitis have been established; all of Koch's postulates have been satisfied in their isolation; however, a specific agent for combatting the poliomyelitis viruses and the method of their transmission are still lacking. The relationship of exertion in the early stages of infection and the severity of subsequent involvement is unquestioned. In the acute stage of the disease, these patients are primarily medical problems. The acute stage is the period of febrile illness; during which time the disease process is still active. This period includes the first 24 to 48 hours that the patient is afebrile. Depending on the location of involvement in the central nervous system, there are three principle forms that poliomyelitis assumes:

1. *The usual case* which may be abortive, non-paralytic, or paralytic.
2. *Bulbar poliomyelitis:* the most severe form in which the virus invades the brain or brain stem. Over half of the patients with bulbar type will have cranial nerve involvement, the tenth cranial nerve being the one most frequently affected.
3. *High spinal thoracic involvement in which the muscles of respiration are paralyzed*, i.e., the intercostal muscles and/or diaphragm.

Because of the endemic and epidemic nature of poliomyelitis and its seasonal incidence, it was felt that an outline of its management in the acute stage would be a timely subject. This management differs depending on the type of involvement which confronts the practitioner. One feature is common to all types of involvement: *exertion is to be avoided at all costs.*

I. The acute stage of spinal poliomyelitis usually includes the first 24–48 hours that patient is afebrile.
   A. Principles:
      1. Avoid exertion; *sacrifice everything for this principle.*
      2. Prevention of deformities.
   B. Treatment:
      1. *Absolute bed rest:* patient is not even permitted to feed himself; he may assume any position which is restful.
      2. Positioning of parts in neutral anatomic positions:
         a. Kenny bed with bed board and foot board.

b. Elevate head of bed 4 inches.
c. Knee roll: knees flexed 45 degrees.
d. Shoulder pads.
e. Hand rolls.

3. Hot packs: many types; all equally effective
   a. Effective in reducing spasm and pain.
   b. Use only if patient is having severe spasm and pain.
   c. Not used more than three or four times daily; ½ hour each session.
   d. Maximum benefit when each packing session is immediately followed by physiotherapy.
   e. Extra salt intake if hot packs are used: Thermotabs, ½ to 1 tablet three times a day.
4. If the patient is restless and has severe pain, sedation with opiates is not contraindicated.
5. Diet: a high caloric and high vitamin intake is recommended.
6. Urinary retention: encountered in significant number of patients in the acute stage.
   a. Treated with FURMETHIDE, a parasympathomimetic drug. Dose: ¼ to 1 cc. subcutaneously, may safely be repeated at ½ hour intervals. Usually complete emptying of the bladder results 15 minutes after administration.
   b. Patient maintained on oral Furmethide, sublingual tablets, 5 mgms., three times a day.
   c. *Caution: Do not use in bulbar patients.*
7. Passive physiotherapy
   a. Early institution is recommended.
   b. Trained nurses are in the best position to administer early passive physiotherapy during their daily ministrations to the patient.

II. Bulbar Poliomyelitis
   A. *The respirator is contraindicated because of:*
      1. The danger of aspiration of secretions.
      2. The inability of bulbar patient to accommodate with respirator.
   B. Trendelenburg position: 8 inch blocks under foot of bed.
      1. Patient permitted to lie on his side or in the prone position, but *not in the supine position.*
      2. Encourage patient to keep oropharynx clear of secretions.
   C. *Absolutely nothing by mouth in acute stage:* danger of vomiting and aspiration.
   D. Suction of the oropharynx whenever necessary.
      1. A soft, rubber catheter is best for this purpose.

2. Never suction patient through the nose.
3. Leave decision to suction up to the nurse.
4. All attempts should be made to secure cooperation of the patient.

E. Check blood pressure and pulse rate at frequent intervals; a rising blood pressure and pulse rate are poor prognostic signs.

F. *Absolutely no sedation:* danger of central depression.

G. Parenteral feeding.
1. Keep daily fluid intake at minimum recommended level, 45–50 cc. per pound per day.
2. High parenteral vitamin intake.
3. High protein intake; the daily administration of blood or plasma in a dose of 10 cc. per pound per injection is probably best for this purpose.

H. Procaine penicillin, 300,000 to 600,000 units daily as a prophylactic measure against secondary invaders.

I. *Do not gavage feed these patients in the acute stage.*

J. Indications for tracheotomy: a tracheotomy set should be kept at the bedside.
1. Inadequate, untrained personnel.
2. Extremely restless or uncooperative patient.
3. Vocal cord paralysis.
4. Bulbo-spinal poliomyelitis.

K. Administration of oxygen.
1. Administered at rate of 8 liters per minute.
2. Best given by nasal catheter.
3. Patient must be watched constantly; for this reason a tent is impractical.

L. Bulbar patients require *constant supervision* and are never to be left alone at any time.

M. A careful check is kept on the respiratory rhythm.
1. *Irregularities of rhythm* imply respiratory center involvement.
2. Consider the possibility of using of the electrophrenic stimulator when such involvement occurs.

III. High Spinal Cord Paralysis: Respirator is life-saving in this group of patients. Respiratory distress due to intercostal and/or diaphragmatic paralysis. Characteristically respiratory rate is rapid but *REGULAR*.

A. Hot packs are placed on the chest and nasal oxygen is administered for ½ hour, if the respiratory distress is not relieved, then the patient is placed in a respirator.

B. The respirator is run at a rate of approximately 20 per minute, with a *negative* pressure of from 10 to 15 mms. maintained.

C. Positive pressure in the respirator is used only for short intervals to encourage coughing. *Do not use positive pressure for prolonged periods of time.*
D. Frequent motion and massage of the patient are mandatory.
F. A high fluid intake is recommended.
G. A low calcium diet should be ordered.
H. *Absolutely no sedation.*
I. The routine administration of prophylactic penicillin in a dose of 300,000 to 600,000 units daily to combat secondary invaders.
J. Positive pressure oxygen by mask or artificial respiration for 15 minutes three times a day to prevent atelectasis is a worthwhile measure.
K. Sluggish bowels are encountered frequently in the respirator patient. Regular bowel pattern should be insured in these patients.
   1. A mild laxative is given each night at bedtime.
   2. Enemata are administered if necessary.
L. Positioning of parts, physiotherapy, and hot packs may be utilized while patient is in the respirator.

JOSEPH M. LOPRESTI, M.D.

CELIAC DISEASE

Special Report No. 237

John J. Calabro, M.D.

INTRODUCTION

Celiac disease is a distinct entity which can be differentiated from all other comparable illnesses by: (1) a characteristic history: (2) a characteristic symptomatology, and (3) a specific response to a definite mode of therapy. No other disease falling under the celiac syndrome can befit such a characteristic three-fold pattern.

The term "celiac syndrome" has been very much abused in recent years. In view of the current opinion[1, 2] that children who were formerly diagnosed as having "starch intolerance" or "infantile steatorrhea" actually represent cases of celiac disease which are mild or which occur under one year of age, a proposed classification of the celiac syndrome follows:

I. celiac disease
II. mucoviscidosis (fibrocystic disease of the pancreas)
III. miscellaneous (allergy, tuberculous peritonitis, chronic enteric infection due to ascaris, salmonella, girardia, etc.)

INCIDENCE

The incidence of celiac disease is difficult to surmise. In the last two decades, there has come a widespread feeling that the disease is actually far more common than earlier estimates would indicate.

Gee has shown that all ages are susceptible but that the peak age falls in the one to five year range, with an onset most frequently during the second year. Celiac disease may exist from birth and is not at all uncommon under one year of age[1, 18]. How frequently celiac disease may occur after age six is difficult to estimate. It has been postulated that with increasing age the disease manifests itself in a somewhat different fashion and with less severity, so that many cases among older persons are not reported[1]. There is some evidence to support the contention that in adults celiac disease is identical with non-tropical sprue. There is no significant difference in incidence among different socio-economic groups or between the sexes.

Statistical data clearly shows that the disease has been found in several generations of the same family, as well as in twins, in siblings, and in cousins[1, 18].

ETIOLOGY

Despite numerous theories (bacterial infection, allergy, hypothyroidism, nervous system disturbance, vitamin deficiency, faulty fat metabolism, endocrine disturbance, general constitutional weakness), the etiology of celiac disease remains obscure[10, 11, 13, 14, 22, 23, 25, 27, 28, 31]. The hypothesis herein presented is that proposed by Anderson and upholds that celiac disease is based on an hereditary constitutional defect capable of manifesting itself only under certain circumstances[1]. Accordingly, the same basic defect can result in different but comparable disease processes depending upon the age and upon the circumstances. Thus, the celiac child may suffer from non-tropical sprue, non-specific colitis, or a mild digestive disorder as an adult. The evidence for a basic constitutional defect is as follows: (1) *As regards siblings:* Severe celiac disease has on occasions occurred among siblings. More striking is the fact that a patient with severe celiac disease commonly has a sibling affected by a mild form. Two or three children with the mild type are commonly found in the same family. Celiac disease has occurred in fraternal twins and in identical twins. (2) *As regards relatives other than siblings:* Several instances of its occurrence in parent and child and in first cousins are well-documented. Members of previous generations of the same family often have digestive or metabolic disorders[14].

The trigger mechanisms which seem to lead to clinical disease are: (1) a period of poor diet (poor in protein, fruits and vitamins but high in

starch and fat), (2) repeated or chronic respiratory infections, (3) enteric infection, (4) emotional tension or trauma.

CLINICAL STAGES AND SYMPTOMATOLOGY

For the sake of convenience, this disease can be divided into three clinical phases: (1) the prodromal (evolutionary) period, (2) the celiac state at which time the classical picture of the disease becomes manifest, and (3) the maintenance phase. In many cases, the classical celiac state does not occur and in these cases, the disease process is very mild and has been properly termed "mild celiac disease"[1]. It is comparable to the entity that has been improperly referred to in the past as "starch intolerance".

(1) *Prodromal (Evolutionary) Stage:*

The disease usually begins insidiously with various digestive disturbances anytime after birth. During this vague evolutionary period, the child may show each of the following: (a) marked anorexia with resultant retardation of growth; (b) changes in behavior and disposition—usually marked as increasing irritability; (c) frequent loose stools and occasionally, vomiting. Periods of constipation may alternate with the periods of diarrhea. Stools at this stage reveal an abnormally high fat content.

(2) *Classical Celiac Stage:*

This represents the full-blown picture of celiac disease and is most frequently encountered during the second year of life. The characteristics ascribable to the classical celiac state follow:

A. *Age of onset* is usually under one year but may be later in breast-fed infants. The first symptom of the celiac triad (diarrhea, poor weight gain and enlarged abdomen) usually begins between 0–9 months. The triad is typically completed by 12–15 months. The diagnosis is never made before three months of age. Vomiting is frequently present.

B. *Body contour* is typical, there being an enlarged abdomen, weak and atrophied muscles, flat buttocks, and scanty subcutaneous tissue. Abdominal enlargement may be absent during periods of anorexia.

C. *Psychological state* is characteristic and the child shows irritability, lethargy, anxiety, and emotional lability. The irritability and anxiety are often shared by other members of the family.

D. *Stools* during a period of normal dietary intake are pale, bulky, and contain an excess of fat, chiefly as soaps, and also of starch and undigested food particles. There is intermittent diarrhea. Although intracellular starch (contained within the membrane of a vegetable cell) may be found in the feces of a normal infant, extra-cellular starch granules are found only in

the feces of patients who lack amylase (i.e., celiacs) and who have been fed cereal starch or whose bowel motility is rapid.

E. *Hypoproteinemia* is found in the majority of patients who have not received a high protein diet (greater than 6 gm. of protein per kilo per day). There is suggestive evidence that the difficulty in protein metabolism shown by the high requirement for dietary protein and a tendency to hypoproteinemia on a normal diet, may be an expression of the basic disease[35].

F. *Intolerance to a normal diet* and clinical response to dietary therapy.

G. *Chronic course* with tendency to relapse, especially when the diet is relaxed or a respiratory infection is present. There is spontaneous improvement during the interval between 3–6 years. Growth and development may be impaired in the more prolonged and poorly-treated cases. Occasionally, dwarfism and even deformity may result[36].

H. *Deficiency of one or more vitamins* may be present. Common deficiencies are scurvy, rickets, tetany, xerophthalmia, tongue lesions, megaloblastic anemia.

I. *Motility of the small intestine* as seen on X-rays after a barium meal shows dilation of the small intestine and excessive segmentation of the barium, with coarsening and obliteration of mucosal folds. Clumping and coarsely granular flocculation of barium are frequently seen. Hypertonicity and hypermotility may be present in less advanced cases.

J. *Pancreatic amylase* is absent or low in nearly all cases[1]. Pancreatic trypsin[4] and lipase[1] are present in all cases[1]. Accordingly, a consistent and objectively demonstrable characteristic of the basic disease is a deficiency of pancreatic amylase, associated with an excess of starch in the feces when starch is fed and clinical intolerance to dietary starch. The assay of amylase is not a reliable means of diagnosis, however, because of its low concentration during the first months of life in normal infants, and in many older infants and young children with chronic diarrhea[2]. Although amylase may occur in patients with pancreatic fibrosis, presumably because of the failure of salivary amylase to be destroyed in passing through the stomach, trypsin is never present—and this represents an important point in the differential diagnosis[3, 4].

(3) *Maintenance Period:*

The maintenance period is prolonged and not without relapses. Normal weight is usually recovered in a few years but the patients may continue to have their brief recurrences of diarrhea precipitated by any of the trigger mechanisms already enumerated. After an interval of one to several years, it is possible to relax the dietary restrictions somewhat without ill effect, and by school age, these restrictions can usually be mild. Late

recurrences (even in adulthood) have been observed and these may respond less readily to treatment.

(4) *Mild Celiac State:*

These cases are far more frequent than the severe cases and show the following characteristics:

(A) *Intermittent diarrhea* beginning during the first year of life. Most attacks are initiated by an upper respiratory infection.

(B) *A high familial incidence.* Several cases may occur in siblings or first cousins. It is not infrequent in siblings of a child with severe classical celiac disease.

(C) *Clinical presence of undigested extracellular starch* in the feces, and low duodenal amylase.

(D) *Clinical response to the diet* which is effective in patients convalescing from severe classical celiac disease.

(E) An enlarged abdomen and flabby but not atrophic muscles, with body contours like those of a patient convalescing from severe classical celiac disease.

(5) *Celiac Crisis:*

The celiac crisis can occur anytime during the celiac state, i.e., before complete recovery. It occurs usually after some mild parenteral or enteral infection or with dietary indiscretion, but occasionally with no obvious cause demonstrable and is characterized by a watery diarrhea and vomiting with resultant dehydration and acidosis. This may be so severe as to result in death if not properly treated.

DIAGNOSIS

The diagnosis of celiac disease depends upon three factors: (1) history; (2) sufficient clinical investigation to rule out the presence of other diseases of specific etiology; and (3) response to appropriate dietary therapy.

(1) *History:*

Whenever an infant or a young child presents a history of two or more episodes of diarrhea, the possibility of celiac disease must be considered. Wide variation exists—from patients exhibiting few to many loose or watery stools for a period of days or weeks—those having multiple intermittent episodes of loose frequent stools. This may alternate with periods of normal stools or even constipation and may finally culminate within a period of months to a state where, although there may be only one or two stools a day, each of these is large, pale, greasy and of foul odor. The history will also show that if the periods of diarrhea have existed for a

sufficiently long period of time, there is evidence of loss of weight, or failure to gain weight, and of failure to grow. The degree of retardation is dependent upon the duration and the severity of the disease. Another important feature in the history is the change in the psychological picture of the patient from a normal child to a depressed youngster who has developed severe behavior problems. Unexplained irritability should make one suspicious of the possibility of celiac disease—if some of the other symptoms are present only to a minor degree.

(2) *Clinical Appraisal:*

As far as clinical investigation is concerned, the picture in the severe case of long duration is clear. Mild cases, however, may appear in an excellent nutritional state.

Examination of the stool will be of assistance primarily in eliminating the possibility of a severe parasitic infestation or bacterial infection of the intestinal tract. The examination of the stool for fat is useful only if the fat content of the feces is found to be markedly elevated[18]. While celiac disease is the principle cause of this condition, it can occur in other diseases as well. In celiac disease the occurrence of steatorrhea is a variable which depends on the severity of the diarrhea at the moment when the specimen for examination is passed[6]. A finding of normal fat content of the stool in no way excludes the diagnosis of celiac disease. The discovery of starch granules in the stool is of dubious significance because their presence depends upon the briskness of the diarrhea at the time of the collection of the sample. If there are frequent loose movements, the intestinal passage time is short, and many items of ingested material may be found in the stool. Moreover, the feeding of banana to a normal child will result in the appearance of many extracellular starch granules in the stool. Therefore, the presence of starch granules in the stool serves to confuse rather than to illuminate the diagnosis of celiac disease[18].

Shwachman stresses that the lysis of gelatin film by a stool indicates the presence of trypsin, and that this test can be used to differentiate cystic fibrosis of the pancreas from celiac disease[32]. However, it should be emphasized that the action on which this test is based cannot be solely attributed to pancreatic trypsin because the stool contains many proteolytic factors, both from the child himself and from the organisms of the intestinal tract. However, since the stools from most cystic fibrosis do not cause lysis of a gelatin film, this test may be a useful screening device to distinguish cystic fibrosis from celiac disease.

The study of duodenal enzymes is fraught with technical difficulties and resultant errors. The most that can be said for this test is that an appreciable trypsin content rules out the possibility of cystic fibrosis[42]

An analysis of the blood sugar is useful in excluding the possibility of diabetes. It is now recognized by most workers that the flat glucose tolerance curves reported in celiac patients merely represent an expression of the poor state of absorption characteristic of the disease during periods of severe diarrhea.[18] Direct and indirect evidence has been produced by Emery to show that children in the celiac state can absorb and utilize glucose normally, but lack available glucose[15]. Emery also suggests that the cold sweating in children with celiac disease may often be due to acute carbohydrate insufficiency independent of the blood sugar level[16].

Pratt et al[30] maintain that pancreatic fibrosis and true celiac disease can be recognized by the presence of an extensive reduction in the ability to absorb vitamin A. This is especially significant if a single sample of blood is taken at an appropriate interval (four hours for infants under 6 months and five hours for older children) after the administration of a standard dose of vitamin A (60,000 U.S.P. Units of vitamin A per gram of body weight)[30]. According to Haas, however, the value of the Vitamin A absorption curve is even less reliable than the flat glucose tolerance curve[18]. In any event, the Vitamin A absorption curve may serve as a good prognostic tool since it becomes normal when the celiac patient recovers from his disease[26].

Intestinal X-ray examination by barium is useful in ruling out the presence of congenital or postnatal anomalies, such as intestinal shunts or obstructive lesions of various sorts. It has now been shown that the intestinal segmentation pattern is characteristic of many vitamin deficiency states and is not delineative of celiac disease[18]. However, if the segmentation pattern exists, it may add somewhat to the diagnosis establishment. A chest roentgenogram is useful in ruling out the possibility of tuberculosis and in helping to differentiate between a celiac disease and cystic fibrosis. Roentgenograms of the bones may reveal such abnormalities as rickets, osteoporosis, and scurvy. The degree of such abnormalities will depend upon the patient's state of malnutrition but will not cast any significant light on the diagnosis of celiac disease per se[18].

Hematological examination will be helpful only in ruling out the possibility of blood diseases or parasitic infestations of the blood stream. The presence or the absence of anemia will depend on the severity and the duration of the malnutrition which is a concomitant of celiac disease. Bacteriologic and serologic studies, especially of stool cultures, are most useful in excluding the presence of specific enteric or other systemic infection.

(3) *Celiac Dietary Therapeutic Test:*

Given a case where the history indicates celiac disease and where the clinical investigation excludes other disease entities, then the diagnosis of

celiac disease can be fully proven by the administration of the celiac diet. The following procedure should be followed: The child should be given a diet which excludes all carbohydrates, except those present in fruits and vegetables, and includes a high protein portion as well as an adequate fat portion. In a period of time that ranges from about 1 to 12 weeks, the diarrhea will clear. Then the child should be given liberal amounts of bread, sugar, potatoes, and other carbohydrates in addition to the prior diet. If the child develops diarrhea again, the diagnosis of celiac disease is absolute —and proper therapy can be instituted.

THERAPY OF CELIAC DISEASE

(1) *Treatment of the "Crisis":*

A celiac crisis with sudden dehydration and acidosis is an acute medical emergency. Upper respiratory infections usually act as the precipitating factor in patients already in unstable balance. Vigorous measures must be taken at once to restore the water and electrolyte balance and to control the infection by antibiotic therapy.

No food is given by mouth until the patient is hydrated; this is usually accomplished in 1–2 days. Water or half-strength saline may be given as tolerated. A protein milk formula with 5 per cent glucose is given at first, providing 30–50 per cent of the caloric requirement. The amounts are increased rapidly and other foods added promptly, full caloric requirements usually being met within a week.

(2) *Dietary Therapy:*

In celiac disease, there is marked intolerance to dietary starch, some intolerance to dietary fat and high requirement for dietary protein. Simple sugars are well tolerated[33]. Because of the malnutrition and fecal loss, a high caloric intake is essential. *Accordingly, the principles of the diet are as follows:*

(a) high caloric value;
(b) high protein content and simple sugars;
(c) restricted starch and moderate fat;
(d) multiple vitamins in liberal amounts;
(e) no raw cow's milk during early years.

Some of the more common errors in therapy are worthy of mention. (a) *Chronic failure to meet patient's caloric needs* can lead to retarded recovery and growth and even death. (b) *Fear of fruits and vegetables* is partly based on the belief that the presence of visible food particles in the feces means clinical intolerance to that food. (c) *Lack of fear of bread and cereals.* A child with celiac disease will tolerate a wide variety of fruits, vegetables and sugars but does not tolerate cereal foods. (d) *Treatment of the acute diarrheal episodes* associated with infection by excessive and prolonged

dietary restriction rather than by antibiotics. (e) *Failure to continue diet for a sufficiently* long period. Each case should be treated individually, but, as a rule, restrictions can be gradually relaxed between three and six years of age and a balanced normal diet may be allowed thereafter.

The course of the disease may be divided into four phases for convenience in the discussion of dietary therapy. This is an arbitrary division. The actual course is continuous and the duration of each phase varies in the individual case. Phase I (Severe Phase) is characterized by irritability, hydrolability, lethargy, steatorrhea, anorexia, diarrhea, and marked wasting. Phase II (Mild or Recovery Phase) is manifested by good appetite and gain in weight but prompt untoward response to normal diet or to infection. Phase III (Maintenance Phase) is prolonged and is delineated by clinical or near-clinical recovery while on an appropriate diet but without chemical recovery. Phase IV (Quiescent Phase) occurs when there is clinical recovery on a normal diet.

In the *Severe Phase*, a no-starch, low-fat, high-protein diet is essential. The formula should provide two-thirds of the calories. Under 18 months, protein milk and glucose or banana powder is given. Over 18 months, powdered or boiled skimmed milk is used in place of protein milk. Infants are kept on this diet regardless of their condition until after 18 months of age. Calories should average 150 per kilo. Protein should be not less than 6–8 gm. per kilo. No raw milk should be given. Some fat is provided in protein milk and egg yolk. Simple sugars make up the remaining calories and can be given as glucose, ripe banana, fruits, fruit juices, honey and corn syrup. Ripe bananas are exceptionally well tolerated[33, 10]. No starch can be given. Vitamins A, D, and C are administered in liberal amounts; oral preparations of B-complex providing more than the estimated requirements are available. Thomson et al. recommend the use of folic acid in the treatment of the anemia[34]. Dalton goes further to claim clinical improvement of three cases of celiac disease with folic acid (and other vitamins) while on a normal diet[12].

In the *Mild Phase*, the formula consists of dried or boiled skimmed milk with added glucose or banana powder. Supplements can be given as in the severe phase. For those under two years of age and for the first few months in older children, pureed fruits and vegetables can be given.

A moderate-starch, moderate-fat diet is given during the *Maintenance Phase*. Additions are made one by one until the child receives one serving (moderate, i.e. 60 gm.) of one type of starch per meal, a small pat of butter or equivalent of fat, and homogenized milk. The following are better tolerated and added first: well-cooked or puffed cereals, rice, angel or sponge cake, and salads. The following are less well-tolerated and are

added later: macaroni, spaghetti, bread, white crackers, pretzels, ice cream and peanut butter.

A normal balanced diet is given during the *quiescent phase.*

PROGNOSIS AND COURSE

Recovery from celiac disease is a long process which is measured in years. It is characterized by periods of rapid improvement and equally rapid remission. Usually, the diarrheal exacerbations are preceded by infection or dietary indiscretion, but at times occur without obvious cause. The length of time during which the disease is recognized and untreated seems to bear a direct relationship to the time required for recovery; one to two years is an average time with present-day therapy[20]. As recovery occurs, the diarrhea disappears first, then the nervous and psychic disturbances. Finally growth in weight and height begins. Strangely enough, high fecal fat may persist for variable periods up to 15 months after the patient's physical status appears normal. This quiescent stage of the disease has been termed latent celiac disease in which clinical recovery precedes biochemical recovery. Complete recovery is said to be had when the celiac patient can pass normal stools on a normal diet without relapse for a period of three years. The studies of May and McCreary indicate that the Vitamin A absorption test may be helpful in following the recovery of the patient[26]. These investigators suggest that when recovery from celiac disease is complete, the absorption of Vitamin A becomes normal. Spontaneous recovery at puberty has also occurred.

Older series held a 10–15 per cent mortality rate, the chief causes of death being intercurrent infection, celiac crisis, and any of the various vitamin deficiencies. Present series give a 2–3 per cent mortality. Of 100 patients handled by Dr. Anderson in the past ten years, only three have died. The causes of death in this series were transfusion accident, virus pneumonia, and chronic failure to provide adequate calories. The reasons for the lowered mortality are antibiotics, greater knowledge of vitamins, greater knowledge of nutritional requirements in Celiac disease, and improved methods in parenteral therapy in dehydration and acidosis.

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RUPTURE OF THE BOWEL IN A NEWBORN INFANT

Case Report No. 238

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Rupture of the bowel in the newborn is an uncommon occurrence attended by an extremely high mortality. It may occur as an intrauterine accident, at birth, or during the first few hours of extrauterine life. Prompt recognition and early surgical intervention can save some of these patients.

The condition was first described in 1825 by Siebold[1], who reported a case of rupture of the stomach in a newborn infant. Since that time single and series of cases have been reported and reviewed. In 1939 Thelander[2] collected 46 cases of rupture of the bowel in the newborn. One of the most comprehensive papers on the subject is that of Lee and MacMillan[3] which appeared in 1950. All authors agree that the incidence is considerably higher than most figures would indicate, since the condition often goes unrecognized and death is attributed to other causes. Autopsies are usually not performed on stillborn or newborn infants, and even when a careful postmortem examination is done, a minute perforation may go unrecognized and death is attributed to a primary peritonitis.

Confusion as to true incidence results from the multiplicity of names attached to this condition, because of variation in the sites of perforation, and different etiologic factors. Some of the terms used to describe this condition are: fetal peritonitis, melena neonatorum, pneumoperitoneum, and meconium peritonitis.

Although it is well recognized that gastric and duodenal ulcers, the

appendix, or a Meckel's diverticulum may rupture in utero or in the first few hours of life, these conditions are excluded from the present discussion. Their cause is not understood, but treatment is the same as when they are encountered later in life.

Only two cases have been reported in which recovery followed intestinal perforation in the newborn. The first of these was a rupture of the terminal ileum with no apparent distal obstruction and recovery followed simple closure of the perforation. The other case represented an atresia of the transverse colon in which the cecum was the site of rupture and recovery followed resection and anastomosis. Single perforation is the usual finding although multiple ones do occur. The site of this rupture varies with the point of obstruction but gastric blowouts are less common than those occurring elsewhere in the gastro-intestinal tract. Thelander[2] found 16 gastric perforations, 30 duodenal, and 30 lower in the intestinal tract.

Thelander[2] states:

> An infant with perforation peritonitis present at birth or occurring soon after presents a fairly characteristic picture. The little patient looks sick. He is cyanotic, respirations are rapid and grunting, the abdomen is distended, and the abdominal wall, the flanks and the scrotum or vulva are usually edematous. Frequently, brawny induration of the edematous area, which may resemble erysipelas, is also present. Food is taken poorly or not at all. Vomiting is frequent and persistent. The vomitus contains bile and may contain blood. The stools are either absent or scant. Some mucus or blood may be passed. The temperature may be subnormal but varying degrees of fever have been reported. The blood count is of little or no value. The hemoglobin content may be very high which probably indicates only dehydration. The leukocytes may or may not respond with a rise. In cases of intrauterine perforation, the abdomen has, at times, been so distended at birth that delivery was impossible until paracentesis of the abdomen had been performed.

When the rupture is due to atresia or meconium ileus; stools, mucus, or blood are not passed.

X-ray of the abdomen will reveal a marked pneumoperitoneum. Air can be seen in the bowel and some loops may be greatly distended. A fluid level will be present only a few hours following perforation due to peritoneal irritation. The use of contrast media such as barium or lipiodol in studying the upper gastro-intestinal tract is extremely dangerous because of the possibility of aspiration in an infant who is already vomiting. Of much more diagnostic importance is a carefully-administered contrast enema which will reveal small, undistended colon and confirms the impression of a complete block above. Lee and MacMillan[3] emphasize the significance of this finding and how erroneous a diagnosis of microcolon would be. Once intestinal continuity is reestablished, the undistended segment of bowel will quickly assume normal proportion.

Rupture of the bowel in the newborn may result from a variety of causes:

1. *Intestinal atresia* is the most common etiologic factor. The complete block increases the pressure above and when distension becomes sufficient to make a portion of the bowel wall ischemic, there is resultant necrosis and perforation. This allows air and intestinal contents to leak out and decompression follows. The blood supply then improves and the area about the perforation may revert to normal. The bowel proximal to the atretic portion is always large; that part distal, small and collapsed. Ladd and Gross[7] state that perforation never occurs from obstruction due to stenosis. The treatment of choice is resection of the atretic area with primary anastomosis. Devitalized bowel, if present, must be resected. Exteriorization of any segment of bowel in the newborn is extremely hazardous and should be avoided. Even though the distal bowel appears minute, it will rapidly approach normal size and function if adequate anastomosis is performed.

2. *Meconium ileus* is second in importance as an etiologic factor. Farber[5] has emphasized that impaired secretion of pancreatic enzymes results in improper digestion of the meconium allowing it to become thick, sticky, and almost tar-like in character. It may become impacted in the bowel—usually the terminal ileum—resulting in complete intestinal obstruction. This condition, which suggests an acute fulminating type of fibrocystic disease of the pancreas, appears to be more common than atresia, but the occurrence of perforation associated with it is less frequent than with atresia. Once considered uniformly fatal, patients with this disease may now be offered a more favorable prognosis as the result of recent investigation. A 5 per-cent saline solution of pancreatin either introduced into the bowel or given by mouth will liquefy the tar-like meconium and enable one to flush out the inspissated material. There has been great progress with the utilization of this powerful enzyme, but it must be used with extreme care because of its corrosive action outside the bowel lumen. Intestinal resection is not indicated unless nonviable bowel is present. The inspissated bile should be extracted and kept in a liquid state with the aid of pancreatin.

3. *Obstetrical trauma* might be suspected of being contributory to perforation in the newborn. Actually, it is seldom a factor and no verified cases are reported. It is difficult to see how small, undistended bowel could be damaged during a delivery which produces a living child. Except in the most unusual cases, air is not present in the intestinal tract until after birth. When abdominal distension is noted at the time of delivery, it results from peritoneal fluid or causes other than bowel distended with air.

4. *Enemas* poorly administered or given by unskilled personnel have been suggested as another etiologic factor. Here again, analysis of cases

fails to confirm this impression, for with careful search other factors are found responsible for the rupture. It is difficult to so increase the intraluminal pressure without a block somewhere in the intestinal tract.

5. *Spontaneous* rupture of the bowel can occur without complete obstruction, but it is an uncommon occurrence. If an associated anomaly is sought for, it usually can be found. It is extremely dangerous to assume that the perforation is a spontaneous one unless all other causes are carefully excluded. This diagnosis should be the last one entertained, but when present, simple closure will suffice. A possible explanation for this entity is considered to be an intrauterine infection or a septicemia occurring at birth.

6. *Other causes* such as *congenital bands* or *volvulus* may be responsible for rupture. Malrotation is usually present with the constricting bands and volvulus often accompanies the obstruction caused by atresia or meconium ileus. The treatment for these conditions is the same as when encountered without perforation.

CASE REPORT B. G. F. 52-3915

This two-day old white female was admitted to Children's Hospital on April 2, 1952 because of vomiting associated with abdominal distension. Delivery, 48 hours prior to admission, was full-term and spontaneous. Hydramnious was noted and delivery was very difficult because the infant's abdomen was so greatly enlarged. Physical examination revealed a normal-appearing infant except for a tremendous abdomen. On catheterization several ounces of urine were obtained following which the abdomen appeared less distended. An enema was given and a small amount of meconium was recovered but this had no effect on the size of the abdomen. Two subsequent catheterizations were productive of moderate amounts of urine, the last specimen of which was cloudy and contained many white blood cells. The abdominal distension increased during the 48 hours of observation. A tube was passed into the stomach but no accumulation of air or fluid was found. When fluids were started by mouth, the child did not tolerate them and vomited several times. Glucose was administered by clysis.

On admission to Children's Hospital, physical examination revealed an acutely ill 48-hour old infant. The lungs were clear; the heart was normal except for a marked tachycardia. The skin was dry, but dehydration was not marked. The entire abdomen was tremendously distended and tympanitic. Rectal examination revealed the rectum to be patent, and a catheter was passed into it for a distance of about 5 inches. Hemoglobin was 16.5 grams; white blood cells numbered 3,900. Urine contained many white blood cells.

Upright x-ray of the abdomen revealed a marked pneumoperitoneum with a fluid level at the umbilicus. Air under the diaphragms separated them from the liver. Small segments of air-containing bowel could be seen throughout the abdomen. It was apparent that an intestinal perforation was present and operation was indicated.

Prior to operation a tube was passed into the stomach and gentle suction started. Chloromycetin and pencillin were administered intramuscularly; intravenous fluids were started and the patient was cross-matched with whole blood.

One hour and a half after admission, the patient was taken to the operating room where, under gas, oxygen and ether anesthesia, an exploratory laporatomy was performed through a right rectus incision. On opening the peritoneum, a large quantity of air under increased pressure was released. This was accompanied by 200 to 300

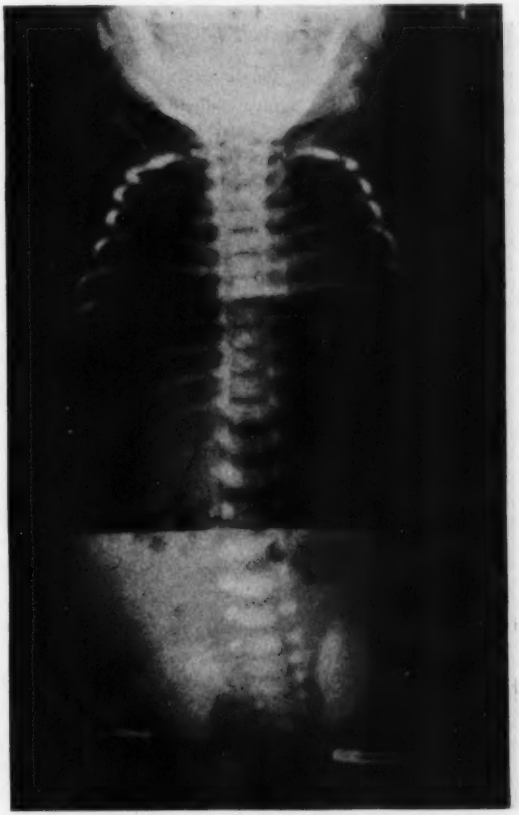

FIG. 1. Erect Roentgenograms revealing marked pneumoperitoneum. The liver has been forced downward and medially by air under the diaphragms. A fluid level is present. Air-containing segments of bowel can be seen throughout the abdomen.

milliliters of thin, cloudy fluid. Once this air and fluid were evacuated, careful exploration was carried out. The intestines were found covered with a thin sheet of whitish-yellow fibrin. There was a perforation about the size of a cigarette tip on the anterior wall of the stomach along the midportion of the greater curvature. The surrounding gastric wall was discolored and the edges were non-viable—characteristic of ischemic necrosis and not ulceration. Loops of bowel were bound down

about this site, and the bladder, which was considerably dilated and injected was attached to these loops in the region of the perforation. Gastric juice, flowing over the dome of the bladder, undoubtedly had caused a chemical cystitis. The cecum and ascending colon were not attached in the usual fashion, but were free with a long mesentery. The small bowel lay in a circular fashion in the midportion in the

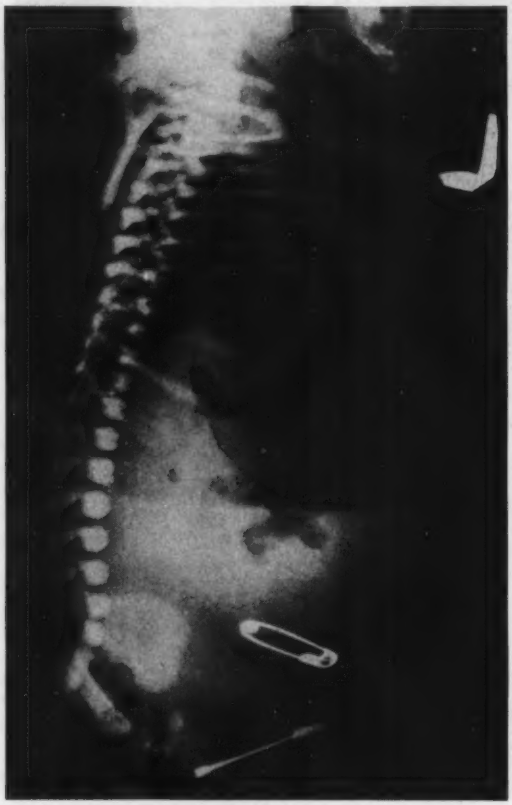

FIG. 2. Lateral roentgenogram shows the same findings as in Fig. 1. The abnormally high position of the diaphragm and the degree of abdominal distension are well demonstrated.

abdomen. No volvulus was present. The right lateral peritoneal reflection was incised and the duodenum was inspected. A search through the small bowel failed to reveal any stenosis, band, or area of atresia. The lower 6-inches of terminal ileum contained black, semi-solid meconium which was tar-like in consistency.

A small rectal tube was passed by an assistant and through it saline was easily introduced up to the ileo-cecal valve. There was no point of contriction although

the calibre of bowel was small. A catheter was passed through the gastric perforation into the duodenum and air and fluid could be inserted with ease down to the point of the meconium impaction. The kidneys, spleen and pancreas were normal in shape, size, and position.

Realizing that this was a meconium ileus with gastric rupture, the cecum was then opened and the meconium was extracted by milking it through the ileo-cecal valve. Ether and saline were injected to aid in this maneuver. Following this, a small catheter was placed through the cecostomy opening into the terminal ileum and was brought out through a small stab wound in the right lower quadrant. The catheter had multiple openings and, when clamped off, acted as a T-tube maintaining intestinal continuity. The gastric perforation was closed with a continuous chromic catgut suture and oversown with interrupted silk sutures. The wound was closed in layers; an indwelling catheter was inserted into the bladder and attached to straight drainage. The child was returned to the ward and placed in a heated crib with continuous oxygen. Intestinal decompression was maintained with Wangensteen suction on the gastric tube.

A 5 per-cent solution of pancreatin was prepared and plans were underway to administer it through the catheter and later by mouth in order to liquefy any remaining meconium.

Penicillin and intramuscular chloromycetin were continued. Whole blood and 5 per-cent glucose were given but the child's condition remained critical. She appeared weaker; respirations became more labored, and eighteen hours following operation, respirations ceased. Permission for an autopsy was refused.

DISCUSSION

This case represents an example of rupture of the stomach due to meconium ileus. The appearance of the perforation and the extent of the peritonitis suggested that the perforation occurred in utero. The degree of abdominal distension, which, undoubtedly, was due to free peritoneal fluid and not intraluminal air, was sufficient to cause dystosia and in our opinion definitely places the time of rupture prior to delivery. The outpouring of acid from the perforated stomach resulted in a chemical cystitis with urinary retention. This confused the clinical picture and the correct diagnosis was delayed because of the recovery of large quantities of urine on catheterization and the inability to reduce the degree of abdominal distension with enemas or gastric intubation.

Realizing the hazards attendant to enterostomy, a tube was inserted into the bowel in such a way that it function as a T-tube through which solutions could be injected, and intestinal continuity would remain intact once it was clamped off or removed. The unattached mesentery of the cecum and ascending colon made the patient a good candidate for a volvulus although this finding was not present at operation.

The use of contrast media such as barium or lipiodol in x-ray examination of the upper intestinal tract was not employed and we consider this procedure dangerous. Air contrast studies are perfectly safe and satisfactory.

Realization of the existence of such an entity, early and accurate diagnosis and prompt surgical intervention combined with meticulous pre- and postoperative care will save some of these infants.

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CLINICO-PATHOLOGICAL CONFERENCE

Directed by: E. Clarence Rice, M.D.

Assisted by: Rugh H. F. Lau, M.D.

By Invitation: John M. Kennelly, Jr., M.D.

Ruth H. F. Lau, M.D.

This one day old white premature male was admitted at 11:30 A.M. on November 30, 1950, and died at 6:12 P.M. on the same day. The patient was born on November 29, 1950 after a normal gestation of eight months. The delivery was aided by a midwife and the infant was seen thirty minutes after delivery by a physician, at which time the child appeared to be doing well. At 11:00 P.M. on the same day, weak grunting respirations were noted. On the following morning a physician was consulted, following which the baby was admitted to the hospital.

The family history revealed that the father was 32 years old and the mother, 23 years of age, both living and well. There were two other siblings living and well. Two years prior to the birth of the patient, the mother gave birth to twins, both of whom died shortly after birth. There was no family history of tuberculosis, diabetes or syphilis.

On physical examination, the patient appeared to be an immature white infant who was weak and lethargic. His weight was 4 pounds, 2

ounces (1.86 kilograms) and the body length was 43 centimeters. The skin was wrinkled and dehydrated. Greenish fluid was expelled from the right nostril during examination.

The respirations were of grunting type. The lungs were clear to auscultation. The heart revealed no murmurs and the rhythm was regular. The abdomen was moderately distended (circumference 26 centimeters), the liver extending 1.5 centimeters below the right costal margin. The spleen was not palpable. No other masses were noted. The rectum was patent, a meconium stool being passed during the examination.

A catheter was passed easily through the mouth into the stomach and bile-stained fluid was aspirated.

The impressions of the examiner were: 1. prematurity; 2. possible intracranial injury. The patient rapidly became worse; respirations were shallow and cyanosis developed. The infant expired at 6:12 p.m. on November 30, the day after birth. The only medication given was 2.4 milligrams of hykinone intramuscularly. An autopsy was performed the following day. No laboratory work had been done prior to death.

DISCUSSION

John M. Kennelly, Jr., M.D.: The pertinent data available indicates that we are considering a premature and newly born cyanotic infant. Green fluid flowed from the nose; the abdomen was found to be distended and respirations were grunting in type. The patient expired 30 hours after delivery. The original clinical impression of the examiner was sound on a percentage basis as the four main causes of death in the newborn in order of frequency are:

1. Premature birth.
2. Birth injury.
3. Congenital malformation.
4. Diseases and infections peculiar to this period of life.

To begin with, this was a premature infant and we know that these children are prone to expire during the first 24–36 hours of life for a number of reasons. In most instances, respiratory failure is the precipitating cause, and at autopsy the most frequent and often the only finding is atelectasis. Aspiration of amniotic fluid and contents may show on microscopic examination. Other causes of death are intracranial hemorrhage and a degree of immaturity incompatible with life in the vital organs. The infant in this case could have expired from any one of these causes, the most likely being atelectasis or intracranial injury.

The next most common cause of death is that of birth injury and here again intracranial hemorrhage ranks high. The symptoms, when present at birth, are often cardiac or respiratory in nature and do not have to be

accompanied by neurologic manifestations. This could account for the picture seen here.

Congenital malformations, diseases and disturbances peculiar to the newborn account for the cause of death in the majority of the remaining cases. The clinical course and manner of death would seem to point to a chest condition in this case and there are several disease entities which must be considered.

A spontaneous pneumothorax or mediastinum may result from trauma or atelectasis. The chest was described as clear to percussion and auscultation but an x-ray would have definitely ruled this out. A large lung cyst would cause respiratory failure and death if not diagnosed and aspirated. Diaphragmatic hernia with the chest cavity filled with abdominal contents may cause cyanosis and death. Atelectasis has already been mentioned on an aspiration basis.

Vascular anomalies such as an aortic ring with pressure on the trachea has been known to be fatal at this age. Grunting respirations are common in this condition.

Tumors and neoplasms large enough to cause pressure symptoms and respiratory failure are also diagnostic possibilities. Duplications of the alimentary tract at any point from the mouth to the anus may be large enough to cause pressure symptoms.

Tracheo-esophageal fistula is another cause of death on a respiratory basis in the newborn. With this condition, a catheter usually meets a blind end. However, in a rare instance the catheter can be passed through the trachea into the stomach and the correct diagnosis can be overlooked.

The infant was moderately distended and greenish fluid was expelled from the nostrils suggesting an obstruction. To postulate death so rapidly on an obstruction basis, one would almost have to have a perforation with peritonitis. A congenital atresia of the intestine at any point below the ampula of vater, a malrotation of the gut, a duplication, a tumor mass, or a meconium ileus all could cause such a picture.

After going over all these conditions in relation to the case at hand, it still is difficult to come to any definite conclusions. The picture appears to point to a chest condition associated with obstructive symptoms referable to the bowel. I believe a diaphragmatic hernia would best fulfill the picture we have here.

PATHOLOGIC DISCUSSION

Ruth H. F. Lau, M.D.: Post-mortem examination revealed an immature infant weighing 1.8 kilograms and measuring 42.5 centimeters. The brain and neck organs showed no remarkable findings. Examination of the thorax revealed the lungs to appear dark red and firmer and smaller than

normal. The combined weight of the lungs was 44.2 grams. There was slight increase of resistance on cutting, the cut surfaces being dark red and firm. No air or blood could be expressed by manual pressure. The pleural surfaces were slightly injected. Ten millimeters of slightly cloudy yellow fluid was present in each pleural cavity.

Histologically, the lung showed alveoli which were packed with fibrinous exudate, desquamated epithelium, macrophages, and all varieties of leukocytes. The picture suggested aspiration pneumonia.

The abdomen was moderately distended. The peritoneal cavity contained 40 centimeters of foul-smelling, dark-green, purulent fecal material. The peritoneal surfaces were injected, smooth, and of a dull red color. The abdominal organs showed evidence of serositis, resulting from the acute generalized peritonitis. The main pathological finding was in the intestinal tract where the bowel had perforated above an atretic area in the ileum. The esophagus, stomach, duodenum, and jejunum were normal.

The proximal part of the ileum was distended with gas and was dilated to 1 centimeter in diameter. A perforation, 3 millimeters in diameter, was found in the posterior wall of the ileum through which the intestinal contents had extravasated. The site of perforation was 80 centimeters proximal to the ileo-cecal valve. Immediately distal to this perforation, the ileum became atretic, being represented by a solid fibrous cord, 1.6 centimeters in length. The remaining portion of the ileum and the large intestine were collapsed, containing small amounts of meconium in which no cornified epithelium was found at post-mortem examination.

On microscopic examination the serosa of the proximal ileum was found to be thickened and infiltrated by polymorphonuclear leukocytes. This infiltration was also found in the tunica propria and the serosa of the large intestine.

Post mortem culture of the heart blood revealed *E. coli* and clostridia. *Streptococcus* fecalis and *E. coli* were isolated from the peritoneal exudate.

The pathological diagnosis was:

1. Atresia of the ileum with perforation.
2. Peritonitis.
3. Bronchopneumonia.
4. Pleural effusion, bilateral.
5. Immaturity.

COMMENTS

E. Clarence Rice, M.D.: Vomiting of green fluid in a newborn infant should always bring to mind the likelihood of congenital atresia of the intestines although it is of relatively rare occurrence.